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Abstracts

Neurosurgery Session 1

Outcomes of Prenatal Surgery (PRENS) Versus (v.) Postnatal Surgery (POSTNS) for Closure of Myelomeningocele (MM) Studying Patients (pts) in the National Spina Bifida Patient Registry (NSBPR)

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Background The Management of MM Study (MOMS) was an RCT of PRENS v. POSTNS that revealed a lower rate of ventriculoperitoneal shunt insertion (VPSI) at 12 months (mos) of age and better motor function at 30 mos in PRENS pts, "even though those in the PRENS group had more severe anatomic levels of lesions" (Adzick et al. NEJM, 2011). Of the 5596 pts enrolled in the NSBPR through 2014, 136 pts had had PRENS. A cohort comparison study of outcomes of PRENS v POSTNS was done.
Methods 122 of the 136 PRENS pts could be matched to 277 POSTNS pts for both age $\pm$ 6 months and for SB Clinic site. Assessments at last clinic visits were used. VPSI status was assessed at $\geqslant$ 12 mos old. Motor function level (MFL) and ambulation status were assessed at $\geqslant$ 5 years old, when the examination becomes robust (McDonald et al. Arch PMR 1986). MFL categories were sacral, low lumbar, mid lumbar, high lumbar, and thoracic. Ambulation status categories were community ambulation, household ambulation, and not ambulatory. Wilcoxon and chi-square tests were used.
Results 99 PRENS pts and 220 matched POSTNS pts were $\geqslant$12 mos old. There was a lower rate of VPSI (54/99; 55%) in PRENS than in PONS pts (180/220; 81%; $p < 0.001$). There was a lower rate of VPSI in PRENS pts from MOMS centers (13/33; 39%) than from other PRENS centers (40/64; 62%; $p = 0.034$). Chiari II surgery was done in 7/122 (6%) PRENS pts v.19/277 (7%) PONS pts ($p =$ ns). 67 PRENS and 155 POSTNS pts were $\geqslant$ 5 years old. There were no differences between groups in distributions of MFL categories ($p =.831$; ns); distributions of ambulation status categories ($p =.192$; ns); nor in rates of spinal cord detethering surgery (29/122; 24% v. 52/277; 19%; $p = 0.28$; ns).
Conclusions We found a lower rate of VPSI in PRENS than in POSTNS. We did not find that PRENS was superior to POSTNS in motor function.

Chiari II Decompression in Myelomeningocele Patients in the National Spina Bifida Patient Registry

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Background The purpose of this study is to quantify the proportion of individuals enrolled in the NSBPR with myelomeningocele who underwent
Chiari II decompression and to identify factors and interventions associated with decompression.
Methods In the NSBPR, we identified all patients with myelomeningocele. We then identified those who had undergone at least one Chiari II decompression. To examine the association between Chiari 2 decompression and several variables (lesion level, age, year of birth, and history of tracheostomy/gastrostomy) we used logistic regression.
Results We identified a total of 4448 patients with myelomeningocele, from 26 member institutions, of whom 407 (9.2%) had undergone a Chiari II decompression. 13 of 407 (5.1%) had no recorded treatment for hydrocephalus at any time; 57 (14.0%)

had undergone either tracheostomy, gastrostomy, or both. Among children who had Chiari 2 decompression, those under 1 year of age were more likely to also have tracheostomy/gastrostomy (RR 2.20 95%CI 1.25–3.90). Logistic regression detected a statistically significant association between the level of the myelomeningocele and Chiari II decompression (thoracic myelomeningocele patients with the highest rate of decompression). The association between functional level and tracheostomy/gastrostomy was not statistically significant. Children born before 2005 were more likely to have Chiari 2 decompression than those born in that year or later (10.1% v. 7.6%, RR 1.32 95% CI 1.08–1.61).

Conclusions There is a statistically significant association between the functional level of myelomeningocele and Chiari 2 decompression surgery. Children under age 1 are more likely to undergo tracheostomy and/ or gastrostomy in addition to Chiari 2 decompression, suggesting that young patients with symptomatic Chiari 2 may have abnormal brainstem function that is not responsive to decompression.

Rate of Shunt Revision as a Function of Age in a Population of Myelomeningocele Patients with Shunted Hydrocephalus

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Background It is generally accepted that cerebrospinal fluid shunts fail most frequently in the first years of life. The purpose of this study is to describe the risk of shunt failure for a given patient age in a cohort with shunted hydrocephalus secondary to myelomeningocele.

Methods We analyzed data from our institutional Spina Bifida (SB) research database including all individuals with myelomeningocele and shunted hydrocephalus. For our entire population, we determined the number of shunt revisions in each year of life. We then calculated the number of patients at risk for shunt revision during each year of life, thus enabling us to calculate the rate of shunt revision per patient in each year of life. In this way, we are able to evaluate the timing of all shunt revision operations for our entire clinic population and evaluate the likelihood of having a shunt revision during each year of life.

Results A total of 655 patients are enrolled in our SB research database. 519 of these have a diagnosis of myelomeningocele. The mean age is 17.48 $\pm$ 11.7 years (median 16, range 0–63 yrs). 417 patients have had a CSF shunt for treatment of hydrocephalus and thus are included in the analysis. There were 94 shunt revisions in the first year of life. This represents a rate of 0.23 revisions per patient in that year. Rate of shunt revision per patient-year decreases with increasing age, with the exception of an increase in frequency in the early teen years. Shunt revisions continued to occur as late as 43 years old.

Conclusions These data substantiate the idea that shunt revision surgeries in myelomeningocele patients are most common in the first year of life. We also observed a persistent risk for failure well into adult life. Our findings underscore the importance of routine follow up of all myelomeningocele patients with shunted hydrocephalus and will aid in counseling patients and families.

Sleep Disordered Breathing in Myelomeningocele Patients

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Background This study is an exploratory analysis of sleep-disordered breathing in children with myelomeningocele and the effects of hydrocephalus management on sleep organization.

Methods We performed a retrospective review of all patients seen in our multi-disciplinary Spina Bifida clinic evaluating polysomnographs performed between March 1999 and July 2013. We analyzed symptoms prompting evaluation, results, and recommended interventions using descriptive statistics. We have also conducted a subset analysis of nine children who had polysomnography both before and after neurosurgical intervention.

Results Of 52 total patients, 81% received a diagnosis of sleep apnea. The most common presenting symptom was "breathing difficulties" (18, 43%). Mild sleep apnea was present in 26 (50%), moderate in 10 (19%), and severe in 6 (12%). Among the 42 patients with abnormal sleep architecture, predominant pattern was obstructive in 30 and central in 12.

In the subset analysis of nine patients who had sleep studies before and after neurosurgery, there was a trend towards decrease in the mean number of respiratory events (from 34.8 to15.9, $p = 0.098$); obstructive

events (14.7 to 13.9, $p = 0.85$); central events (20.1 to 2.25, $p = 0.15$); and apnea- hypopnea index (5.05 to 2.03, $p = 0.038$).
Conclusions A large proportion of children with myelomeningocele undergoing polysomnography showed evidence of disordered sleep on initial sleep study. Furthermore, 31% had moderate or severe obstructive sleep apnea. Myelomeningocele patients with abnormal sleep structure who underwent non-operative treatment with peripheral oxygen supplementation showed improvement in the apnea-hypopnea index. Our results suggest that polysomnography in myelomeningocele may present an opportunity to detect and classify sleep apnea, identify low risk interventions, and prevent future implications of sleep-disordered breathing.

Practice Preferences for Neurosurgical Management in Spina Bifida: a Survey of the American Society for Pediatric Neurosurgery- Part I: Neonatal and Infancy Issues/Challenges
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Background Despite advances in neurosurgical care there appear to be significant disparities in approach to common problems. To better quantify these preferences we developed a survey that widely explored common Neurosurgical issues in patients with Spina Bifida and utilized Survey Monkey to distribute it to 232 members of the American Society of Pediatric Neurosurgery (ASPN).
Methods A broad based survey was developed by a neurosurgical working group that widely surveyed practice preferences encountered across the lifespan in patients with SB. The survey was distributed via Survey Monkey. There were 32 ASPN members who were not available by e mail or had retired from clinical practice. Of the remaining 200 members there were responses from 80 members (40% response rate). All results are self -reported, non-validated responses.
Results The majority (58%) of centers surveyed closed between 5–15 new open MMC/yr. Approximately 38% of closures occur within 0–24 hours following birth and 60% occur between 24–48 hours of life. Plastic surgery consultation is routine for 12% of centers and never occurs in 20% with the remainder requesting Plastic Surgery assistance on an as needed basis. On occasion 45% of ASPN members support resection of the placode. Only 30% temporize hydrocephalus at the time of back closure. Pre-closure screening test preferences will be discussed. Stridor and progressive ventricular enlargement are the most important two findings to motivate treatment of hydrocephalus in infancy.
Conclusions While significant disparities exist surrounding key decision making for the newborn or infant with MMC there are central areas of agreement that appear supported by solid majorities of senior academic Pediatric Neurosurgeons.

Practice Preferences for Neurosurgical Management in Spina Bifida: a Survey of the American Society for Pediatric Neurosurgery-Part II: Issues of Childhood through Transition
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Background To better quantify the difference is Neurosurgical practice preferences we developed a survey that widely explored common Neurosurgery issues in Spina Bifida and utilized Survey Monkey to distribute it to 232 members of the American Society of Pediatric Neurosurgery (ASPN).
Methods A broad based survey was developed by a neurosurgical working group that widely surveyed practice preferences for a variety of
Neurosurgical problems encountered across the lifespan in patients with SB. The survey was distributed via Survey Monkey. There were 32 ASPN members who were not available by e mail or had retired from clinical practice. Of the remaining 200 members there were responses from 80 members (40% response rate). All results are self-reported and non-validated.
Results Eighty percent of ASPN surveyed Neurosurgeons obtain routine brain imaging on patients with SB. Symptoms of shunt failure without radiographic change prompt revision in about 1-25% of cases. This cohort was more willing to perform shunt revision for symptoms alone compared with images alone. For an intact shunt with increased ventricles and no symptoms 60% of respondents would observe while 25%

would revise. An asymptomatic broken shunt without ventricular enlargement produced evenly divided responses between observation, intervention and further investigation. Operative shunt exploration is always performed before Chiari II decompression (C2MD) in 56% and performed sometimes in 40%.
Conclusions This cohort of academic pediatric neurosurgeons emphasized symptoms above imaging changes in making shunt revision decisions. The role for shunt exploration before C2MD is widely but not universally embraced. Criteria that cross thresholds to trigger C2MD and tethered spinal cord release (TSCR) will be reviewed.

Practice Preferences for Neurosurgical Management in Spina Bifida: a Survey of the American Society for Pediatric Neurosurgery-Part III: Neurosurgical Perspective on Transitional Care

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Background To better quantify the difference is Neurosurgical practice preferences we developed a survey that widely explored common Neurosurgery issues in Spina Bifida and utilized Survey Monkey to distribute it to 232 members of the American Society of Pediatric Neurosurgery (ASPN).
Methods A broad based survey was developed by a neurosurgical working group that widely surveyed practice preferences for a variety of Neurosurgical problems encountered across the lifespan in patients with SB. The survey was distributed via Survey Monkey. There were 32 ASPN members who were not available by e mail or had retired from clinical practice. Of the remaining 200 members there were responses from 80 members (40% response rate). All results are self-reported and non-validated.
Results A multi-disciplinary SB clinic (MDSBC) is present at 80% of centers. More than 50% of the MDSBCs had specialists in Neurosurgery, Orthopedics, Urology, Physical Therapy, Social Work, Orthotics, Physical Medicine and Wheel Chair Repair. Between 20–50% of clinics have specialists in Developmental Pediatrics, Gastroenterology, Nutrition, Neurology and OB -GYN while small numbers of clinics have additional medical specialty services (Nephrology, Endocrinology) available in the MDSBC. About 70% of clinics staffed by these Pediatric Neurosurgeons are purely pediatric while 30% serve children and adults. A transition program is in effect in 37% of these clinics and and 37% of surveyed Pediatric Neurosurgeons follow their SB patients through adulthood.
Conclusions Transition remains an acute need in coordinated care for SB. About a third of current MDSBCs staffed by this cohort of academic
Pediatric Neurosurgeons follow adult patients and just over a third of Neurosurgeons follow their Pediatric patients with SB into adulthood.

Hydrocephalus in patients with myelomeningocele in the National Spina Bifida Patient Registry

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Background In patients with myelomeningocele, rates of hydrocephalus vary widely. This study seeks to determine the rate of hydrocephalus in patients with myelomeningocele in the NSBPR. We explore the variation in shunting rates across institutions and the relationship between hydrocephalus and the level of the myelomeningocele.
Methods We queried the NSBPR to identify all patients with myelomeningoceles. Patients were identified as having treated hydrocephalus if they had at least one hydrocephalus-related operation. We calculated the proportion of patients enrolled at each institution with treated hydrocephalus. Logistic regression was performed to analyze the relationship between hydrocephalus and functional level. We compared patients born before 2005 to those born later.
Results A total of 4448 patients with myelomeningocele were identified from 26 institutions, of which 3558 patients (79.99%) had undergone at least one hydrocephalus operation. The rate of treated hydrocephalus ranged from 72% to 96% among institutions enrolling more than 10 patients. There was a significant association between the level of the myelomeningocele and hydrocephalus. Compared to patients with sacral level myelomeningocele, those with thoracic or lumbar level had higher rates of hydrocephalus (OR sacral v. thoracic: 0.155, 95% CI 0.112 to 0.215). Patients born

before 2005 had a higher rate of hydrocephalus treatment compared to younger patients (82.1% vs. 76.4%, OR 1.42, 95% CI 1.22 to 1.65).

Conclusions The rate of hydrocephalus treatment in patients with myelomeningocele in the NSBPR is consistent with previously published literature. Our data demonstrate a clear association between functional level of the myelomeningocele and the need for CSF diversion. We observe a decrease in the rate of hydrocephalus treatment in patients born after 2005. This may be a sign of changing thresholds for hydrocephalus treatment in this population.